

Anaesthetic Management of a Patient with Myasthenia Gravis Posted for Tonsillectomy- A Case Report

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ABSTRACT

Myasthenia gravis is a chronic autoimmune disease of neuromuscular junction which causes skeletal muscle weakness and fatigability, characterised by decrement in postsynaptic acetylcholine receptor at neuromuscular junction caused by auto-antibodies destruction. There are three types of myasthenia gravis in children- transient neonatal, congenital and juvenile. Juvenile Myasthenia Gravis (JMG) is an autoimmune disorder that can affect all the skeletal muscles including extraocular muscles, which may lead to fatigability and generalised weakness. This report is about a case of seven-year-old patient weighing 25 kg presented with throat pain since one and half month. Patient was a known case of congenital myasthenia gravis and diagnosis was confirmed by Edrophonium test. He underwent tonsillectomy and was successfully managed under general anaesthesia with no muscle relaxant technique. Tonsillectomy in paediatric patients is not an uncommon surgical procedure. In spite of being a common surgery, it is still challenging to the surgeon as well as the anaesthesiologist as there is a shared airway between the two. Also, in the postoperative period there are chances of post-tonsillectomy bleeding causing airway obstruction, if not diagnosed and treated early could risk the life of the patient. Therefore, this surgery poses an increased risk of mortality and morbidity.

Keywords: Anaesthesia, Autoimmune disorder, Chronic tonsillitis, Congenital

CASE REPORT

A seven-year-old patient weighing 25 kg, presented with throat pain since one and half months. He was a known case of congenital myasthenia gravis and developed drooping of eyelids at age of three years. Diagnosis was confirmed by Edrophonium test. Patient was posted for Tonsillectomy in a known case of myasthenia gravis which is well controlled with drug therapy. Acetylcholine receptor antibody titre was 0.03 nmol/L. Patient was on Tab Pyridostigmine 30 mg twice a day and myasthenic symptoms were under control and continued preoperatively.

In preanaesthetic evaluation blood investigations (haemogram, renal function test, liver function test, serum electrolytes, coagulation profile), electrocardiogram, chest X-ray, airway assessment were done. All were found to be within normal limits. After preoxygenating with 100% oxygen for five minutes and giving premedication (inj. Glycopyrrolate 0.1 mg and inj. Ondansetron 1 mg IV) patient was induced with inj. Fentanyl 25 mcg and inj. Propofol 90 mg. Right nasal intubation was done with flexometallic cuffed endotracheal tube no. 5 without use of muscle relaxant [Table/Fig-1]. Controlled ventilation was continued with 50% nitrous oxide in oxygen with sevoflurane and continuous propofol infusion. Patient was haemodynamically stable intraoperatively. After completion of surgery, patient was extubated safely with adequate tidal volume and intact reflexes. Patient was shifted to paediatric Intensive Care Unit (ICU) for postoperative observation. Patient was followed-up after two days of discharged without any discomfort and was found normal with his all routine continued drug treatment.

DISCUSSION

Myasthenia gravis is a chronic autoimmune disorder which is characterised by decrement in postsynaptic acetylcholine receptor at neuromuscular junction leading to skeletal muscles weakness and fatigability, caused by auto-antibodies destruction or inactivation that decreases neuromuscular endplate capacity to transmit the nerve signal [1,2]. Anaesthesia with myasthenia gravis could be given with or without muscle relaxants, where response to muscle relaxant is



[Table/Fig-1]: Patient posted for surgery.

unpredictable. Patient may be resistant to succinylcholine which is depolarising muscle relaxant. As there are less number of receptors available in patients with myasthenia gravis, they are resistant to depolarising muscle relaxant like succinylcholine, meanwhile they

are sensitive to non depolarising muscle relaxant like vecuronium, atracurium, rocuronium etc., [3].

Neuromuscular weakness and easy fatigability are the characteristics of myasthenia gravis which are due to autoimmune damage to the postsynaptic nicotinic receptors. Many other autoimmune diseases can also be associated with myasthenia gravis like hyperthyroidism, rheumatoid arthritis, systemic lupus erythematosus, pernicious anaemia, thymus hyperplasia and thymoma [4]. Muscle relaxants are avoided and potent inhalational agents are used along with intravenous agents like Thiopentone and Propofol to acquire advantage of both and minimising side-effects like hypotension with combining the drug to facilitate tracheal intubation and provide relaxation for surgery. These patients are usually chronically hypovolemic and vasodilated; they are prone to developing exaggerated hypotension during induction of anaesthesia [5]. Potent inhalation agents allow neuromuscular transmission to recover and as they have low blood solubility they are rapidly eliminated at the end of surgery. Sevoflurane is choice of inhalational agent here not only because of its pleasant induction properties favourable for paediatric patients, but also because there are lesser incidences of excitatory airway reflexes at the time of induction with sevoflurane [6]. Kiran U et al., have used Sevoflurane (MAC 0.5-0.7) as the sole anaesthetic agent for a trans sternal thymectomy [7]. There is controversy for residual neuromuscular blockade reversal when muscle relaxant drugs are used. It is difficult to differentiate muscle weakness in case of cholinergic crisis due to inadequate neuromuscular transmission because of presence of anticholinestrases and antimuscaranics [7]. Gag reflex is often absent and thus such patients are at higher risk for aspiration of oral secretions. The patient's ability to generate adequate ventilation and to clear bronchial secretions is of utmost concern in the recovery room [8]. It is recently found that in normal patients there can be normalisation of tetanic response of peripheral nerve but there are chances that the muscles necessary to protect the airway like neck and pharyngeal muscles are still partially paralysed [5]. Authors present working concept of myasthenia gravis is that

of an acquired immune complex disorder at the neuromuscular junction, impairing neuromuscular transmission in voluntary striated muscle. It is associated with a break in immunologic tolerance with blocking and degrading of acetylcholine receptors. There is widening of the synaptic cleft, associated with partial destruction, simplification and shortening of the postjunctional membrane. Hence, use of muscle relaxants can be deleterious in such patients [9]. The different response of peripheral versus bulbar muscles may be more evident in myasthenic patients and it is preferable to keep intubated especially when oral surgery has been done.

CONCLUSION(S)

Anaesthesia with myasthenia gravis could be given with muscle relaxants, where response to muscle relaxant is unpredictable. So, with this case report authors concluded that patients with myasthenia gravis can be successfully managed by inhalational and IV anaesthetic agents by avoiding muscle relaxants and its side-effects.

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